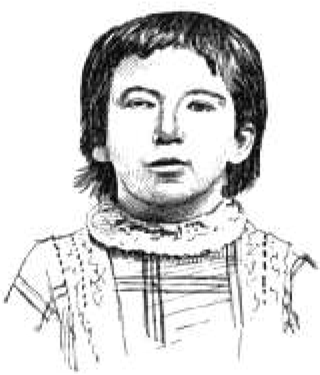
CONGENITAL BILATERAL PLEUROPLEGIA AND FACIAL PARALYSIS.[1]

By A. SCHAPRINGER, M.D.,

NEW YORK.

BECKY F., æt. eight years, was brought to the Eastern Dispensary of this city by her fourteen-year-old sister, on October 11, 1889, because of a cough that was accompanied by a muco-purulent expectorate, and was particularly troublesome at night. She was directed to the deparment for diseases of children, but by mistake went to my department, that for diseases of the eyes. Thus it happened that she came first under my observation. The peculiar appearance of her face impressed me at once and lead mę to make a careful examination, the results of which are detailed below.



Dr. H. Koplik, the attending physician to the department for the diseases of children at the dispensary, at my request, kindly turned the case over to me, and for this great favor I here again take occasion to thank him.

---

[1] Case presented at the meeting of the New York Neurological Society, December 3, 1889.

The disease of the respiratory tract for which the little patient sought medical aid stood in such distant relationship to the remarkable conditions which are described below, that I shall pass it over without any further reference.

The child is of a development corresponding to her age. The deposit of subcutaneous adipose tissue is rather scanty. She is possessed of the average intelligence. The circumstances of the family have been such as not to have permitted them to send her to school, and as a consequence she has not yet learned to read. When walking, she stumbles and falls more frequently than other children of the same age. She declares positively that this is because of a weakness in her legs and not on account of a lack of appreciation of the irregularities and obstacles on the surface of the floor or sidewalk.

She generally holds her head rigidly erect. The glabella is bulging, but otherwise there is nothing unusual about the skull. Her face is pale, expressionless, and presents a mask-like appearance. The naso-labial fold is absent on both sides. There is not the slightest indication of folds in the integument of the face either in laughing or in crying. On one occasion, when she was giving utterance to some peculiar inarticulate sounds, I inquired why she was crying, and was immediately enlightened as to the nature of the sounds by her elder sister, she informing me that the little one was laughing and not crying as I had supposed. The patient cannot close her lips, and in speaking, substitutes linguals for labials, saying, for example, "tata" for "papa," and "nana" for "mamma." The right angle of her mouth is drawn downwards and outwards, and is the only place where any mimetical movements are noticeable. The little expression there is is to be perceived principally when she is talking. Whistling, blowing, etc., are absolutely impossible.

She cannot wrinkle her forehead, either vertically or transversely. When her eyes are open the ordinary and normal amount of eyeball is uncovered. Usually both eyes are open to an equal extent, but quite often one lid or the other hangs a little lower than its fellow. When required, the eyes can be fully and completely opened, showing that the levatores palpebrarum superiorum are not involved. Complete closure of the eyes is impossible, although upon a forcible effort the lids may be so nearly approximated that only a very inconsiderable interval is left between them. To this condition of the lids may be attributed the fact that the eyes have been protected from irritation. There is on either side, quite prominent, that abnormal crescentric fold of skin, covering the inner angle of the eye, and, at v. Ammon's suggestion, called the epicanthus. The caruncle and plica semilunaris are only slightly developed on both sides. They cannot be seen on a front view, and only become visible when looking from the side of the face towards the inner angle.

Ordinarily the axes of the eyes are parallel. Frequently, however, the right eye is directed a little more upwards and slightly outwards. If an object be held in the median plane a few feet in front of the child's head and then she be asked to fix her eyes upon it, she does so without difficulty. She is also able to follow the object with both eyes if it be lowered or elevated in the median line. If the object be approached to her face, keeping it in the sagittal plane, she follows it readily; and she will keep her eyes fixed upon a finger held near the nose for a longer period without fatigue than is generally borne normally. These, however, are all the movements of which the eyeball is capable. They are rotation upwards, downwards, and convergence. When an object is moved to either side of the median line and the child is told to look at it, she invariably answers, "I see it;" but she stares directly ahead, turning her eyes neither towards the right nor the left. This same condition obtains if the eyes be alternately covered by the hand. For example, if the right eye be covered and the object then moved out of the median line to the left, the uncovered eye does not follow the object, showing a paralysis of the external rectus of the left side. On the other hand, if the object be moved to the right, the eye does not follow it, showing a paralysis of the internal rectus of the same side.

As we have seen, the two internal recti contract freely if required, in order to bring the eyes into convergence for near objects. If, however, one of these muscles be required to act in unison with the external rectus of the opposite eye, for a conjugate lateral version there will be found an utter inability to do so. The two external recti muscles are either completely paralyzed or entirely absent, as neither the right nor the left eye can be directed externally from the median line the least particle. On the other hand, the internal recti are only relatively paralyzed. They converge the eyes readily when required, but they are utterly unable to respond to an impulse which under normal condition would induce a conjugate lateral version. When the child wishes to see an object situated to the side of the median plane, instead of rotating her eyes she turns her entire head.

Under normal conditions the range of binocular fixation (not to be confounded with the field of vision) may be said to correspond to a geometrical solid with three dimensions. In the case under consideration, however, the range of binocular fixation is reduced to a sagittally placed plane, and as such it has but two dimensions, one of which is vertical and the other sagittal.

With the ophthalmoscope the vessels in the fundus appear to be more tortuous than usual. Otherwise nothing worthy of mention is to be seen.

The left eye is practically emmetropic, having only a slight degree of astigmatism. The right eye is myopic to the extent of about 3 D. This is the eye which, as was stated before, is directed upwards and slightly outwards when looking at a distance. In order to avoid losing the good will of the child I refrained from carrying the rather wearying tests for the determination of the acuity of vision out to completeness. As far as could be determined, V. was nearly, if not quite, normal in both eyes.

The ophthalmoscopical observations were conducted without the aid of mydiatics, and the little patient's efforts of accommodation proved a great obstacle. They showed, however, that the functions of the ciliary muscles were unaffected, which could also be demonstrated in other ways.

With reference to certain analogous cases reported by other observers, it would be well to state explicitly that there is no strabismus convergens, and that there is not the least trace or scar of a former tenotomy or myotomy, and that, furthermore, in looking downwards there is no abnormal convergence of the axes of the eyes.

The size and mobility of the pupils are perfectly normal.

When the tongue is protruded it is deflected a little to the left of the median line. The left half of the tongue is a trifle smaller than the right half.

Movements may be made in all directions, but not with the requisite completeness and force. When eating, the child is obliged to use her finger to dislodge food from her cheeks. She says that she is unable to masticate hard substances,such as bread-crusts. If a hard body is placed between her teeth, she is able to hold it there quite firmly. She can move the lower jaw laterally towards the right side, but not towards the left, indicating a paralysis of the right external pterygoid muscle. She has uvula bifida. The child herself called my attention to a remarkable anomaly in her mouth, which I should certainly not have thought of looking for otherwise. The cord-like structure, which may be felt by passing the finger along the buccal mucous membrane between the alveolar process of the upper jaw and the cheek in the neighborhood of the canine fossa, and spoken of by the dentists as the lateral *frenulum* or canine ligament, is enormously developed in this case on the right side, while on the left side the state of its development is normal. What significance can be attached to this condition I am not prepared to say, my opportunities for making anatomical investigations in regard to it having been too meagre. Hearing, taste and smell are normal, as is also the sensibility of the skin.

I wish to express my great obligations to Dr. Geo. W. Jacoby for helping me during the examination of the nervous symptoms of this case.

The distal phalanx of the left index finger is bent out of the long axis of the finger at an angle of about 150° in the direction of the middle finger. The affected interphalangeal joint possesses the normal mobility, and there is not the least trace of a previous inflammation. All the circumstances point towards the defect being a congenital one.

The anterior aspect of the thorax shows an extraordinary deformity. The sternum is deeply sunken in, and presents a long vertical groove situated beetween two prominences formed by the bulging of the anterior portions of the sites on either side. Dr. Abraham Jacobi, who examined this deformity at my request, pronounced it a not very well-marked example of that form of imperfect development known in the literature of the subject as "funnel chest."

No other deformities, such as webbed fingers and toes are discoverable.

The parents of our patient are natives of Russia, where the patient was also born. They are divorced and live in distant parts, while the child and her elder sister are being brought up by an uncle. The parents are not consanguineous. The sister of whom mention was made before is physically well developed and looks healthy. Another child, however, of the same parents died at the age of a few months. The child who is the subject of this paper is said by her relatives to have been free from any anomalies at birth, except the before-mentioned angular index finger. The relatives also state that the distortion of the mouth was developed subsequent to an attack of convulsions which occurred in her early infancy. According to another version, the blame attaches to injury received by striking her head in a fall. It is perfetly manifest that no weight can be given to those accounts, and that we must consider the anomalies as congenital.

The most interesting features of the case are unquestionably the lesions of the nervous system. They include the motor branch of the trigeminus, the hypoglossus and the facial, but particularly those nerve tracts of both eyeballs that control the conjugate lateral version. I should like to confine myself to the consideration of this last anomaly entirely.

Previous to this there has been no simple and concise term to designate the condition of paralysis of the conjugate lateral version of both eyes to the right or to the left, or, as in this case, to the right and the left with preservation of convergence. I therefore suggest the name "Pleuroplegia," deriving it from the Greek word *pleuron*, the original meaning of which is "the side." The word is constructed similarly to "Pleurothotonus," by which we designate a convulsion that draws the body to one side and is seen in tetanus. In the succeeding portions of this article the reasons for the introduction of this kind of term will be shown to be timely as well as conducive to a clearer conception of the subject.

Pleuroplegia (or to be more exact, ophthalmopleuroplegia,) is generally seen in diseases of the pons, such as neoplasms, hæmorrhages, the softening due to arterial thrombosis, etc., and is usually unilateral. In attempting to explain this phenomenon we have to reason about as follows:

Given a case where there is left-sided pleuroplegia, with the power of convergence retained, the following parts must be undisturbed in regard to their anatomical structure and their functions: 1st. The two internal recti muscles. 2d. That branch of the motor-oculi that supplies these muscles. 3d. The two nuclei for the internal recti. 4th. The two intracerebral paths which connect the nuclei of the internal recti with the centre of voluntary convergence. This is supposed to be a single centre situated in the cortex. 5th and last. This convergence centre itself. In our supposed case the lateral conjugate movements toward the left side are completely arrested. This movement is accomplished by the external rectus of the left eye and the internal rectus of the right eye.

We shall now study the innervation of these individual muscles separately.

Since the left external rectus is completely paralyzed, the following series of neuro-muscular structures may be regarded as having been rendered inactive: 1st, the left rectus externus; 2d, its supplying abducent nerve; 3d, the abducent nucleus in the pons; 4th, the nerve tract which connects this nucleus with that portion of the cortex of the right side of the brain in which the centre that presides over the voluntary left lateral version of both eyes is situated; 5th, this centre itself.

The right internal rectus muscle is only relatively paralyzed, that is only in regard to conjugate lateral version. There is no impairment of its power of convergence. If we now follow the chain that connects the right internal rectus with the voluntary centre of conjugate left lateral motion, we have: 1st, the muscle under consideration; 2d, its innervating branch from the right motor-oculi; 3d, the right internal nucleus. At the internal nucleus the impulses of convergence and of conjugate lateral version separate. For the internal nucleus has two lines of communicution with the cerebrum. The one connecting it with the centre of voluntary convergence has been discussed, the other which will now engage our attention is that which connects it with the centre of voluntary conjugate lateral version. This centre is located in the cortex of the right hemisphere, that is on the same side as that internal nucleus to which we had come in the course of our progress from without inwards. The connecting path between the right internal nucleus and the centre of voluntary conjugate lateral version in the cortex of the right hemisphere is not, however, a perfectly straight one confining itself within the boundaries of the right cerebral hemisphere, but takes part of its course through the substance of the left hemisphere, making a sort of loop and passing quite close to the nucleus of the left abducens. This is the same abducens nucleus that we met before in the course of our analysis. So near to this nucleus do the connecting fibres pass that a lesion of the nucleus almost invariably involves them. The course of the fibres carrying the impulses of conjugate left lateral motion are undivided from their origin in the cortex of right hemisphere until they reach the neighborhood of the left abducens nucleus. Here, however, they split into two divisions, one of which joins the left abducen nerve, while the other passes over to the opposite side into the right internal nucleus and by means of this communicates with its respective branch of the oculo-motor nerve. If the central communicating fibres are injured in any part of their course, there will result as a consequence an absolute paralysis of the left external rectus and a relative paralysis of the right internal rectus. In other words, a left-sided pleuroplegia. This would, of course, also follow from lesions involving either of the two terminals, that situated in the right cortex, and the left abducens nucleus.

What has been said in reference to left-sided pleuroplegia holds good *mutatis mutandis* of that of the right side.

That the term "paralysis of the abducens" for this condition is insufficient and consequently conveys an erroneous impression, is manifest, but still it is used in the literature of the subject. Perhaps the term "nuclear abducens paralysis" would be morre corect. Even this, however, will not answer as a designation for the clinical symptom as such, because it makes a topical diagnosis, which, while it may apply to a large number of the cases, will not apply to all of them.

The terms "deviation conjuguée" and "paralysie conjuguée" remind us that we are indebted to a group of French investigators for the original clinical observations concerning this symptom. I shall only mention the discoveries of Achille Foville and Feréol. Foville first published, in 1858, the observation that in unilateral disease of the pons there is a paralysis of associated lateral motion of the eyeball toward the side on which the lesion of the pons was situated.

In 1872, Féréol called attention to the seeming paradoxical circumstance that the internal rectus responded freely to the impulses of convergence while it was powerless to carry on the motions of conjugate lateral version. The terms "conjugate deviation" and "conjugate paralysis" are both applicable to disturbances of motion in every possible direction, and it is always necessary to denote the direction, whether upwards or downwards or to the left, etc., by the appropriate words. Of all the conjugate paralyses that which we have called "pleuroplegia" is of the most frequent occurrence, and, because of the twofold rôle played by the internal rectus muscle, the most interesting.

In not every case where pleuroplegia or pleuroparesis exists is the condition to be discovered by any marked deviation of the eyes. This is well illustrated by the following case which came under my own observation :

A four-year-old girl fell down stairs, and was brought to the Eastern Dispensary. Dr. W. W. Van Arsdale, to whom she was first presented, asked me to examine her left ear from which there had been some bleeding. At the time we saw her the child was perfectly conscious and in good spirits. There was no eye symptom complained of, and nothing remarkable was to be seen. It chanced that I had had a case, a short time previously, of paralysis of certain muscles of the eye, and that led me to investigate the mobility of the eyeball in this case. The child when requested to follow a finger with her eyes, could only do so in the left half of the field of fixation. To the right she could only follow a trifle beyond the middle line, and there the eyes remained in a sort of nystagmus-like motion. There was a difference in the two eyes, inasmuch as the left eye went a little further beyond the middle line than the right. I know nothing of the future of the case, as the parents never brought her again.

For the sake of completeness it may be mentioned that the opposite condition to pleuroplegia, namely, a paralysis of convergence, with retained conjugate lateral motion, has been observed and is being regarded with increased attention in the newer literature. If a short expressive term for this symptom is desired, that of "mesoplegia" is suggested.

In the case described here the symptoms are evidently due to a lack of development involving the nuclei of the abducens, facial, hypoglossal and trigeminus nerves.

It is because of the close neighborhood of these nuclei that this seems the most reasonable explanation of the symptoms. The inconstant strabismus sursum vergens, alternately now in one eye and then in the other, denotes that along with the formative defect there is an active unstable cause at work, in regard to whose nature I may be excused from theorizing.

Only four parallel cases have as yet been recorded in literature. They are the cases of "congenital bilateral abducens facial paralysis" of Alfred Graefe (Halle),[2] G. C. Harlan (Philadelphia),[3] Julian J. Chisolm (Baltimore),[4] and P. J. Möbins (Leipzig).[5] The present one is the fifth case.

Of the five cases under consideration, although agreeing in the principal points, not a single one is exactly similar to any of the others in all of the particulars. In Graefe's case the facial paralysis was complete only on the left side; on the right side there was paresis. In the cases of Harlan, Chisolm and Möbius the paralysis was complete only in the upper portions of the face. In the lower portion there was some mobility about the angles of the mouth. In our case only the right corner of the mouth is mobile.

In regard to the impaired mobility of the eyes, there was in Harlan's and Chisolm's cases congenital converging strabismus. In Chisolm's case the patient had undergone a myotomy of the internal recti in early childhood. When Chisolm saw this patient she was thirty-five years of age, and her eyes were parallel. As in the other three cases, there was in those of Harlan and Chisolm a complete paralysis of the abducens on both sides. These observers have not stated clearly whether the shortened internal recti muscles responded to an impulse of accommodation. Harlan merely says that the function of all the orbital muscles but the abducent seemed normal. Chisolm states that in his case the lateral motion in both directions was absolutely impossible, because the abducens of either side was paralyzed from birth on the one hand, while on the other hand the power of the internal muscles had been completely arrested by the operation for the strabismus. One misses in the last case a definite statement as to the ability to converge, especially as Chisolm found it necessary to order convex glasses for hyperopia, in order to enable her to see clearly while sewing. In the three other cases, namely, those observed by Graefe, Möbius and myself, no converging strabismus was present.

---

[2] Graefe-Saemisch, Handbuch der Augenheilkunde, Bd. vi., p. 60; also Siebenter Periodischer Internat. Ophthalmologen-Congress, 1888, p. 30.

[3] Transactions of the American Ophthalm. Society, 1881, p. 216.

[4] Arch. of Ophthalm., edited by Knapp and Schweigger, vol. xi., p. 323 (1887).

[5] Münchener Medicinische Wochenschrift, 1888, No. 6 (February 7).

There is a remarkable similarity between Graefe's case and the one described in the beginning of this article in regard to the differences of refraction existing in the two eyes. In both cases the right eye is myopic while the left one is emmetropic. Furthermore, in both cases there is the slight vertical deviation of one eye. In Graefe's case, however, the left eye was directed a trifle lower permanently, while in our case the deviation was not constantly manifested, sometimes the right eye and at other times the left squinting upwards. In looking at an object held close to the face, this upward deviation, when existing, would always disappear in our case.

Möbius observed a difference in this respect only when the glance was directed downwards. The mobility of the eyeballs was limited in the downward movement and convergence always took place. It may be well to state here that in our case there was no abnormal limitation of downward movements, and that in fixing the gaze upon near objects in a downward direction there never is an excessive or abnormal convergence.

So far there has not been a report of an autopsy on a case where this complication of symptoms existed.

The epicanthus, the bulging glabella, the uvula bifida, the crooked index finger and the undeveloped funnel-chest are unique accompaniments, as are also the anomalies existing in the regions supplied by the trifacial and hypoglossal nerves. Möbins observed in his case a web between two fingers of the right hand.

The first observation of paralysis of the lateral motion of both eyes was made by Stellwag v. Carion.[6] In his case, however, the affection was not congenital. It occurred when the patient was twenty years of age, in the course of a strumous exophthalmia. The description of the earlier stage of this case corresponds essentially to the conditions found by Graefe, Möbius and myself. The axes of both eyes were parallel to each other and to the median plane, while movements to the right or left were impossible to the slightest extent. The power of convergence was fully retained. The conditions present in a subsequent stage, however, resembled rather those found by Chisolm and Harlan. Both eyes, especially the left one, were directed inwards. If the object the patient was looking at was moved out of the median plane to the right or the left, the corresponding eye only followed it until it stood parallel to the median plane, beyond which it never went, the opposite eye remaining in a strongly adducted position. Later again, the right eye returned to its normal position, while the left eye was directed a trifle to the inner side. During the course of the illness other severe complications arose, such as asthmatic attacks and mental disturbances. The limitations of the mobility of the eyes decreased constantly and the condition of the visual organs had almost returned to the normal state when the patient withdrew from observation.

---

[6] Ueber gewisse Innervationsstörungen bei der Basedow's chen Krankheit. Wiener Med. Jahrb., Bd. xvii., p. 25 (1869). First Case.

A NEW MECHANICAL TREATMENT FOR TABES.

The "Times and Register," Nov. 3, 1889, reports from the "B. klin. Wochenschrift," according to Juergensen, favorable results obtained in tabes dorsalis from the application of Hessing's corsets, made from finely woven cloth and thin steel ribs. The freeing of the spine produced by wearing the corset soon results in relief from cystic and intestinal trouble; then the lancinating pains cease, and the patient walks better. The patellar reflexes are not restored. General improvement is most marked. The corset must be worn for years, under the supervision of the maker. No explanation is given by Juergensen, unless the circulation and nutrition of the cord are improved by the extension of the spine. Erb has seen favorable results in two cases from this treatment. L. F. B.